Autoimmune Hepatitis Accompanied by Idiopathic Thrombocytopenic Purpura and Sjögren's Syndrome

Key words: PAIgG, Shirmer's test, liver biopsy, HLA typing

We report our experience with a 71-year-old male patient, similar to the case reported by Wada et al in the April 2001 issue of this journal (1). Although our patient had been healthy until the end of the previous year, he was referred to Kakogawa Municipal Hospital because of right lower abdominal pain on January 2, 2001. His relevant family history showed nothing noteworthy. On admission, physical examination revealed several petechiae on both lower extremities, while cholelithiasis and a normal-sized spleen were detected by abdominal sonography. Laboratory tests showed a platelet count of $8 \times 10^9/l$, white blood count of $4.6 \times 10^9/l$, and hemoglobin level of 13.6 g/dl. Serum aspartate aminotransferase was 123 IU/l (normal: 8 to 40), alanine aminotransferase 213 IU/l (normal: 5 to 35), lactate dehydrogenase 215 IU/l (normal: 105 to 210), and alkaline phosphatase 381 IU/l (normal: 130 to 330). Results of the coagulation function tests were within normal limits. Test results for surface antigen of hepatitis B virus and anti-hepatitis C virus antibody (Ab) were negative. Immunological studies disclosed IgG of 3,250 mg/dl, IgA of 393 mg/dl, IgM of 443 mg/dl, anti-nuclear Ab= $\times 640$ (homogenous and speckled pattern), anti-DNA Ab= $\times 80$, anti-smooth muscle Ab= $\times 80$, anti-SS-A Ab= $\times 16$, anti-SS-B Ab= $\times 8$, and platelet-associated IgG (PAIgG) of $12,260 \text{ ng}/10^7$ platelets.

Bone marrow aspiration showed normoplastic marrow with a megakaryocyte count of $31/\mu\text{l}$. Results for Shirmer's test (5 mm for the right and 3 mm for the left eye) and for the rose bengal test were positive, and sialography of the parotid gland revealed an irregular conduit pattern. On the basis of these results, the patient was suspected to have autoimmune hepatitis

(AIH), idiopathic thrombocytopenic purpura (ITP) and Sjögren's syndrome (SjS).

Oral administration of 30 mg of prednisolone was started on January 15, 2001, and the platelet count increased to $88 \times 10^9/l$ by February 2 and the abnormal liver function findings also returned to within the normal range.

A liver biopsy on February 1 revealed the presence of chronic hepatitis accompanied by piecemeal necrosis. The HLA typing pattern was: HLA-A2, A11, B54 (22), B61 (40), DR4. These findings led to a diagnosis of definite AIH (18 points), based on the diagnostic criteria proposed by the international AIH group in 1993 (2).

Although Katayama et al (3) reported a case of AIH complicated with SjS and thrombocytopenia, which was caused by antiphospholipid Ab syndrome, as AIH complicated with ITP and SjS is rare, there is a clear need for accumulating clinical experience.

Noritatsu YAMAIKE, Katsuyasu SAIGO*, Shion IMOTO**, Hideyuki MIYACHI, Shiho MORITA, Yuichiro MAEDA, Yoshinobu TOMOFUJI and Tadanobu CHINZEI

The Department of Medicine, Kakogawa Municipal Hospital, Hyogo, *Blood Transfusion Division, Kobe University Hospital, Kobe and **Hyogo Red Cross Blood Center, Hyogo

Reprint requests should be addressed to Dr. Katsuyasu Saigo, Blood Transfusion Division, Kobe University Hospital, 7 Kusunoki-cho, Chuo-ku, Kobe 650-0017

References

- 1) Wada M, Kamimoto H, Park SY, Shintani S, Nakasho K. Autoimmune hepatitis concomitant with hypergammaglobulinemic purpura, immune thrombocytopenia, and Sjögren's syndrome. *Intern Med* 40: 308-311, 2001.
- 2) Johnson PJ, McFarlane IG. Meeting report: international autoimmune hepatitis group. *Hepatology* 18: 998-1005, 1993.
- 3) Katayama Y, Kohriyama K, Kirizuka K, Nishizaki H, Fujii H, Tanji Y. Sjögren's syndrome complicated with autoimmune hepatitis and antiphospholipid antibody syndrome. *Intern Med* 39: 73-76, 2000.